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NIH Fiche to Paper Journal

TITLE:	INSTRUCTIONAL COURSE LECTURES
PUBLISHER/PLACE:	American Academy Of Orthopaedic Surgeons Park Ridge Il
VOLUME/ISSUE/PAGES:	1996;45():425-46 425-46
DATE:	1996
AUTHOR OF ARTICLE:	Gitelis S; Wilkins R; Conrad EU
TITLE OF ARTICLE:	Benign bone tumors.
ISSN:	0065-6895
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CHAPTER 45

Benign Bone Tumors

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Benign tumors of bone represent a diverse group of pathologic and clinical entities. They vary greatly in aggressiveness and clinical behavior, thus requiring a broad spectrum of treatment. Many of these lesions can be observed without any form of intervention; others require complete en bloc excision followed by complex reconstruction.[1] Some of these lesions also possess the potential for malignant transformation followed by metastasis and represent the most serious type of benign bone lesion.[2] Those metastases, while appearing benign histologically, can be lethal. For example, pulmonary metastases can be lethal because of extensive involvement of the lung parenchyma. It is important for the treating physician to understand these wide variations of behavior in order to manage patients properly.

The diagnostic strategies for benign bone tumors center on the initial radiographic presentation.[3] Radiographs in two planes are the best method for evaluating any bone lesion (Fig. 1). Initially, one must determine whether the tumor appears benign radiographically, using several parameters. The first step is to determine what the tumor is doing to the bone. A benign tumor generally is confined by a natural barrier and, while it may expand the bone, it usually does not destroy the cortex and extend into the soft tissue. The second step is to determine how the bone is reacting to the tumor. A benign lesion usually is slow-growing; this allows the bone to marginate, or react to, the neoplastic process. A dense sclerotic margin around the tumor is a characteristic sign of a benign bone tumor. Similarly, if the periosteum has had an opportunity to react to expansion of the bone by forming mature bone, a benign process is suggested. The third step is to determine whether periosteal responses are present. If the tumor creates a sequential layering of periosteal new bone on its surface (so-called onion-skinning), this suggests a rapidly evolving process, which can occur in both benign and malignant conditions. Codman's triangle represents rapid periosteal elevation with reactive changes and is another sign of an active or, in many instances, a malignant process. A sunburst appearance is another radiographic feature of malignancy. Fourth, any extension into soft tissue is an ominous sign and suggests a malignant or very rapidly growing benign process. Fifth, the radiographs are used to determine whether there is any matrix mineralization within the tumor. If the tumor is characterized by destruction of bone, then the presence of calcification or ossification will suggest the type of neoplastic process present. For example, a calcified, lytic phalangeal lesion strongly suggests the presence of cartilage that is consistent with enchondroma. Sixth, the margin between the tumor and the host bone also should be characterized radiographically. Most benign tumors have a geographic type of bone destruction with a sharp zone of demarcation between the tumor and the host bone. On the other hand, permeative destruction represents a gradual zone of transition and is more common in malignant tumors. Finally, the location of the tumor within the bone also is helpful in identifying which type of tumor likely is present. Most benign tumors are metaphyseal; however, giant cell tumor and chondroblastoma typically are epiphyseal.[4] Diaphyseal tumors are rare and include fibrous dysplasia and eosinophilic granuloma. Certain benign tumors, such as aneurysmal bone cyst and osteoblastoma, are located more commonly in the spine, especially in the posterior elements. Cortical locations are more common for osteoid osteoma and infections

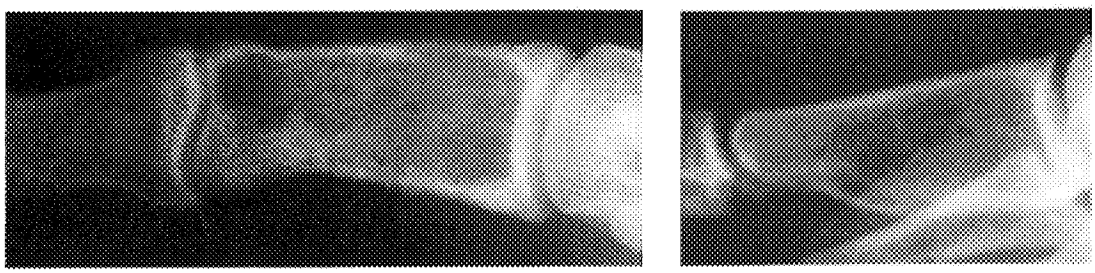

Fig. 1 Left, AP and **right,** lateral radiographs of the proximal phalanx of the great toe of a patient who had a benign aneurysmal bone cyst. Note the geographic type of bone destruction. The bone is expanded but the lesion is marginated by cortical bone.

of bone. Tumors located on the surface of the bone include three benign lesions: periosteal chondroma, periosteal desmoid, and periosteal aneurysmal bone cyst.

In addition to plain radiographs, other studies are important in evaluating benign bone tumors (Fig. 2). Radioisotope imaging with technetium-99 diphosphonate is very helpful in determining whether the process is monostotic or polyostotic. Certain benign tumors, such as multiple hereditary exostoses, enchondromatosis, and fibrous dysplasia, can be polyostotic. A technetium bone scan also is very useful in identifying osteoid osteoma. This lesion often takes up the radioisotope intensely, and the scan is useful in identifying the location of the nidus as well as in showing the adequacy of surgical excision. Three-dimensional imaging of a benign bone tumor is quite useful at times. This can be accomplished with either computed tomography (CT) or magnetic resonance imaging (MRI). For example, CT is very useful in identifying the location of osteoid osteoma. The images created on tomographic scans appear similar to the face of a clock; positioning of the nidus on this clock-face greatly facilitates the surgical approach. CT also is very helpful when approaching a giant cell tumor. It is particularly useful in identifying cortical destruction, which represents the best site for entry into the bone when an intralesional excision is

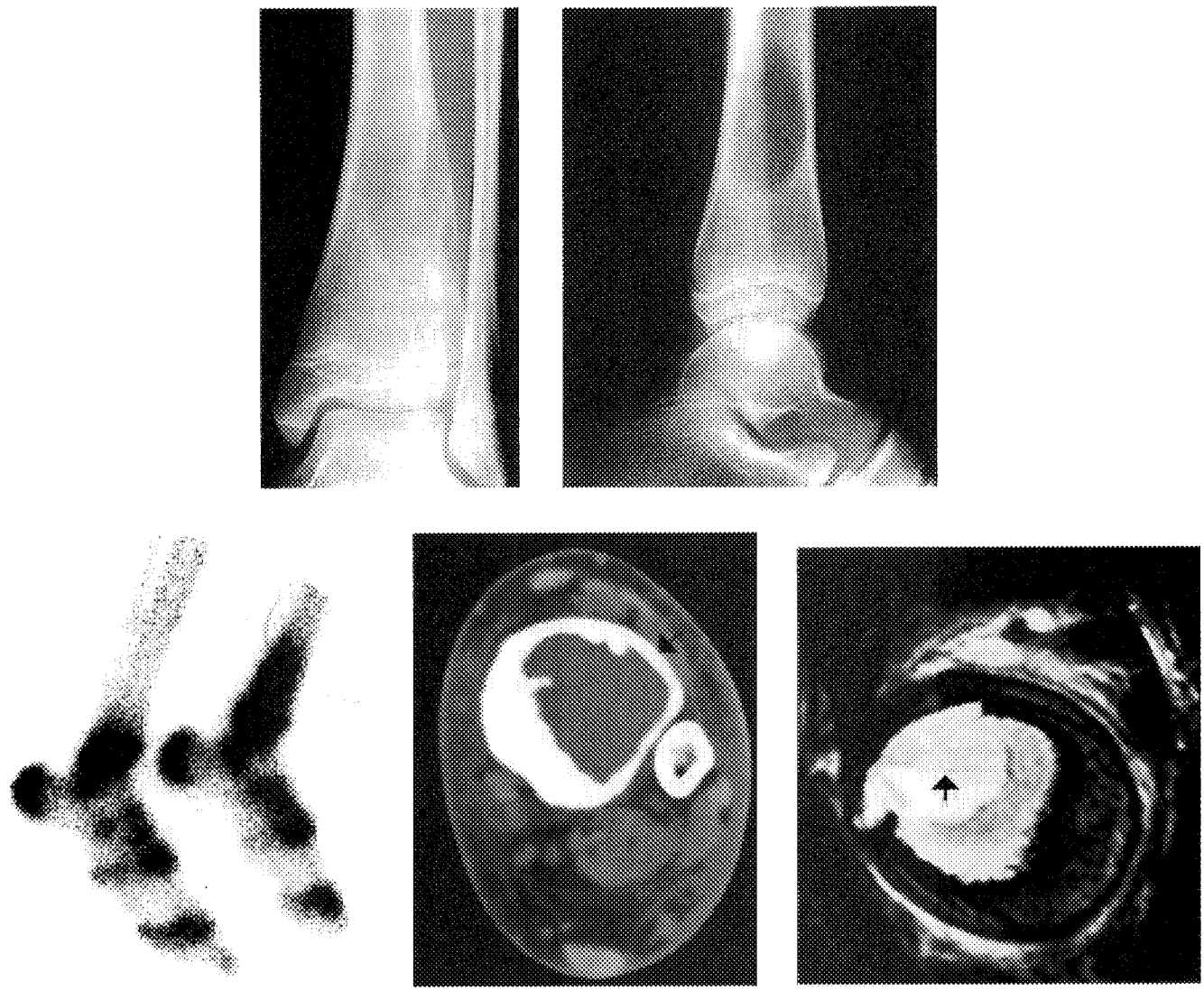

Fig. 2 Top left, anteroposterior and **top right,** lateral radiographs of the distal tibia of a skeletally immature patient with a benign aneurysmal bone cyst.. Metaphyseal bone destruction with benign so-called onion-skin periosteal response is evident. **Bottom left,** Technetium bone scan revealing activity in the lesion compared with the contralateral side. **Bottom center,** CT scan demonstrating containment of the tumor but with cortical thinning (arrow). **Bottom right,** MRI of the tumor. Note the fluid level (arrow) visible on this transverse image.

performed. Identification of the neurovascular bundle is very important when a giant cell tumor extends into the popliteal fossa; this identification is best accomplished with MRI. MRI also is useful in determining the coronal and sagittal extension of a tumor within the medullary canal.

After adequate imaging of the tumor has been performed, the next step in diagnosis is biopsy with excisional, open incisional, or needle techniques.[1,3,5,6] Excisional biopsy is performed more commonly for benign bone tumors than for malignant ones. Rarely is an excisional biopsy done for a malignant tumor. Lesions such as a giant cell tumor in an expendable bone or portion of bone, such as the proximal end of the fibula, are best treated with excision of the involved bone as both a diagnostic and a therapeutic procedure. Other expendable bones include ribs, portions of the scapula, small tubular bones of the foot, and the anterior arch of the pelvis. Benign tumors in these locations can be removed entirely at the time of the biopsy.

Open incisional biopsy is another technique that frequently is used for benign bone tumors. This must be done carefully. The biopsy should be placed in an anatomic site that will allow for the definitive surgical procedure. In addition, meticulous hemostasis must be obtained to avoid creating a postoperative hematoma. The risk of postoperative hematoma can be eliminated by doing an open incisional biopsy followed by intraoperative examination of a frozen section, and then doing the definitive procedure immediately. Finally, needle biopsy is useful for some benign bone tumors. This is especially true for anatomically inaccessible lesions within the pelvis and spine (Fig. 3). Guidance with CT can be very helpful in placing the needle in the lesion.

Nomenclature

Benign tumors of bone generally are classified by the type of neoplastic tissue within the lesion. Seven general categories of tumors are described in the classification system of the World Health Organization (Outline 1).[3,7] Group I includes bone-forming tumors such as osteoma, osteoid osteoma, and osteoblastoma. Group II includes cartilage-forming tumors such as chondroma (enchondroma and periosteal chondroma), osteochondroma (solitary or multiple), chondroblastoma, and chondromyxoid fibroma. Group III includes giant cell tumors such as osteoclastoma. Group IV includes marrow tumors; no benign marrow tumors are included in this classification. Group V includes vascular tumors such as hemangioma, lymphangioma, and glomus tumor. Group VI includes other connective-tissue tumors such as desmoplastic fibroma, lipoma, and benign fibrous histiocytoma. Group VII includes other tumors such as neurilemmoma and neurofibroma.

In addition to these tumors that are classified by the World Health Organization, there are several tumor-like lesions of bone that closely resemble benign bone tumors and are included in this discussion. These lesions include solitary bone cyst, aneurysmal bone cyst, metaphyseal fibrous defect (nonossifying fibroma), eosinophilic granuloma, fibrous and osteofibrous

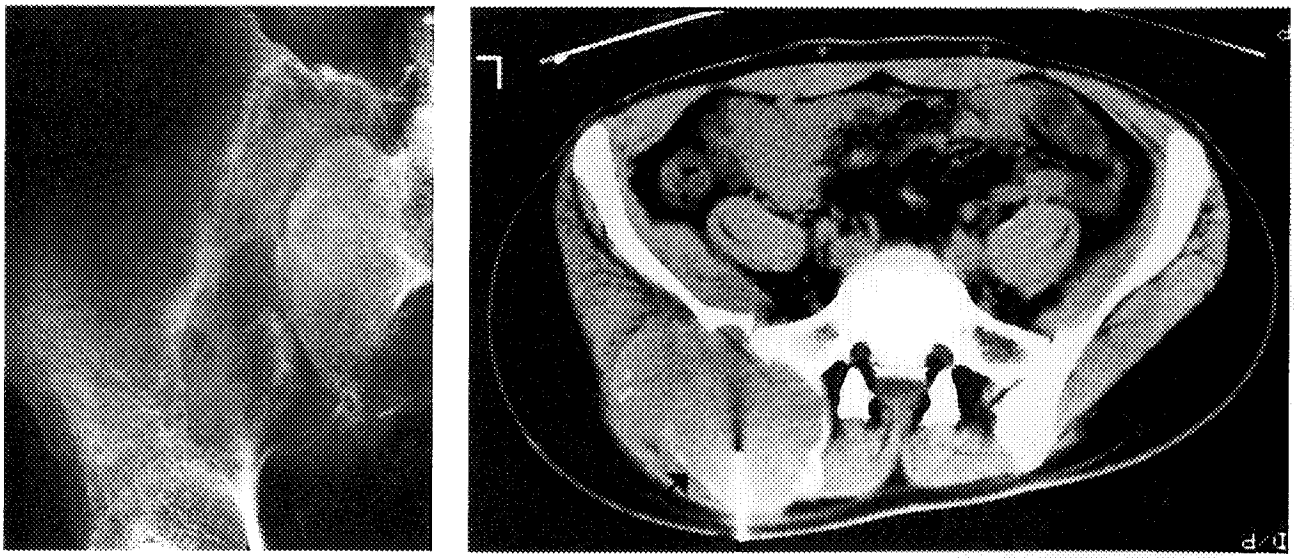

Fig. 3 Left, AP radiograph of a portion of the pelvis of a patient with a giant cell tumor in the pelvis shows a destructive tumor near the sacroiliac joint. **Right,** CT scan of the pelvis shows a large extrapelvic soft-tissue mass (arrow) that proved to be a giant cell tumor.

Outline 1
Classification of benign bone tumors

I. Bone-forming tumors
  a. Osteoma
  b. Osteoid osteoma
  c. Osteoblastoma
II. Cartilage-forming tumors
  a. Chondroma
  b. Osteochondroma
  c. Chondroblastoma
  d. Chondromyxoid fibroma
III. Giant cell tumors
IV. Marrow tumors (none)
V. Vascular tumors
  a. Hemangioma
  b. Lymphangioma
  c. Glomus tumor
VI. Other connective-tissue tumors
  a. Desmoplastic fibroma
  b. Lipoma
  c. Fibrous histiocytoma
VII. Other tumors
  a. Neurilemmoma
  b. Neurofibroma
VIII. Unclassified tumors (none)
IX. Tumor-like lesions
  a. Solitary bone cyst
  b. Aneurysmal bone cyst
  c. Metaphyseal fibrous defect
  d. Eosinophilic granuloma
  e. Fibrous dysplasia
  f. Osteofibrous dysplasia
  g. Myositis ossificans
  h. Brown tumor of hyperparathyroidism
  i. Intraosseous epidermoid cyst
  j. Giant cell (reparative) granuloma

(Adapted with permission from Schajowicz F, Ackerman LV, Sissons HA, et al (eds): *Histologic Typing of Bone Tumors.* Geneva, Switzerland, World Health Organization, 1972.)

dysplasia, myositis ossificans, brown tumor of hyperparathyroidism, intraosseous epidermoid cyst, and giant cell (reparative) granuloma.

Finally, several syndromes are associated with polyostotic disease. For example, polyostotic fibrous dysplasia is associated with several endocrine abnormalities under the eponym of Albright syndrome. Eosinophilic granuloma is called Hand-Schüller-Christian disease when associated with exophthalmus and diabetes insipidus, and it is called the potentially fatal Letterer-Siwe disease when associated with involvement of organs such as the liver and spleen. Enchondromatosis may be polyostotic (Ollier disease), and it may be associated with vascular abnormalities under the eponym of Maffucci syndrome.

Staging

The most widely accepted staging system for benign and malignant bone tumors was devised by Enneking and associates[1,8] at the University of Florida (Table 1). Three stages of benign tumors are defined by Arabic numerals. Stage 1 is a latent benign bone tumor, stage 2 is an active benign bone tumor, and stage 3 is an aggressive benign bone tumor. A latent (stage-1) lesion is one that does not progress or that heals spontaneously. An

Table 1
Staging of benign bone tumors[1,8]

Stage	Type
Stage 1 (latent)	Nonossifying fibroma
	Enchondroma
	Unicameral bone cyst
	Osteochondroma
	Osteoid osteoma
	Fibrous dysplasia
	Eosinophilic granuloma
Stage 2 (active)	Enchondroma
	Osteochondroma
	Osteoid osteoma
	Osteoblastoma
	Giant cell tumor
	Chondromyxoid fibroma
	Fibrous dysplasia
	Eosinophilic granuloma
	Aneurysmal bone cyst
	Unicameral bone cyst
	Osteofibrous dysplasia
Stage 3 (aggressive)	Giant cell tumor
	Osteoblastoma
	Chondroblastoma
	Aneurysmal bone cyst

example of a latent benign lesion of bone is a nonossifying fibroma (Fig. 4). This lesion frequently is picked up as an incidental finding on a radiograph, and there have been many examples of spontaneous healing. An active (stage-2) lesion is one that can expand and even deform the bone but that is fully contained by the bone (Fig. 5). Giant cell tumor of bone frequently is a stage-2 lesion. A stage-3 lesion is one that invades and destroys the bone and extends into the soft tissues. This stage does not refer to metastasis, histologic appearance, or rate of growth. Giant cell tumor, osteoblastoma, and chondroblastoma can behave in this manner and can be so destructive that they mimic a malignant tumor

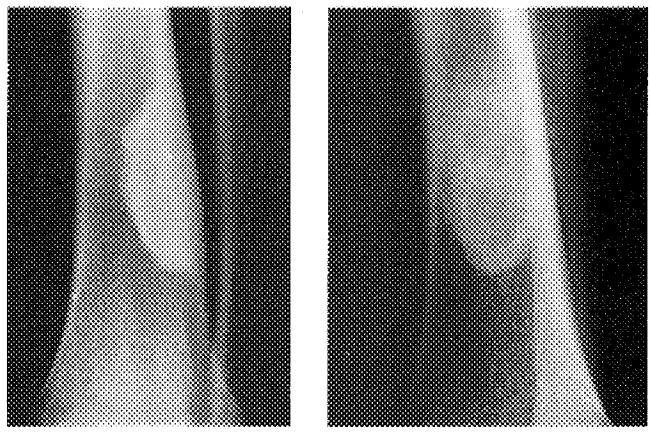

Fig. 4 AP, **left**, and lateral, **right**, radiographs of the distal tibia, showing an ossified eccentric metaphyseal lesion that is a healed nonossifying fibroma. This is a latent (stage-1) lesion.

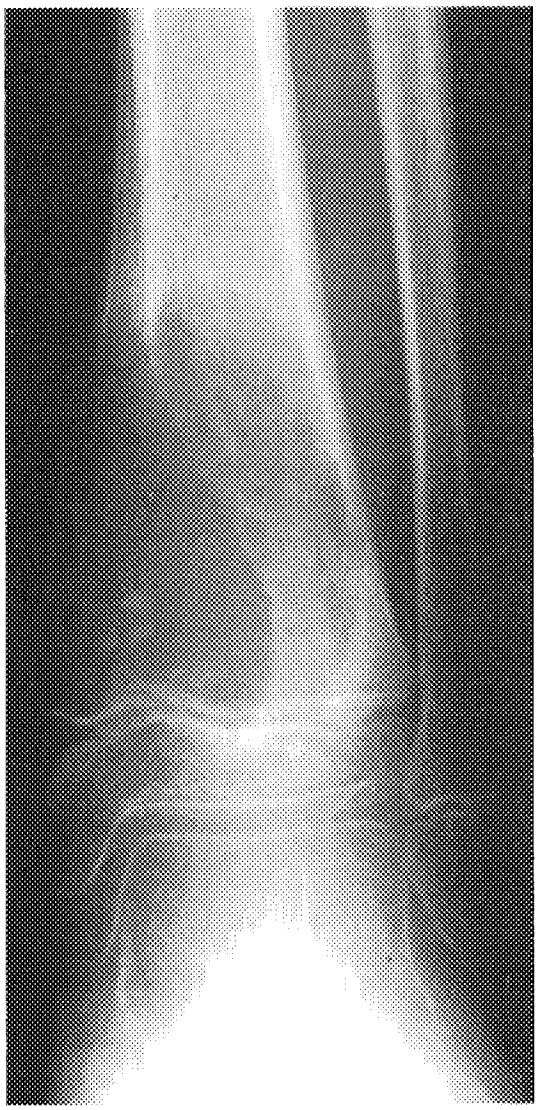

Fig. 5 AP radiograph of the distal tibia of a child with an active (stage-2) aneurysmal bone cyst. The metaphysis is expanded but contained by a thin cortical rim.

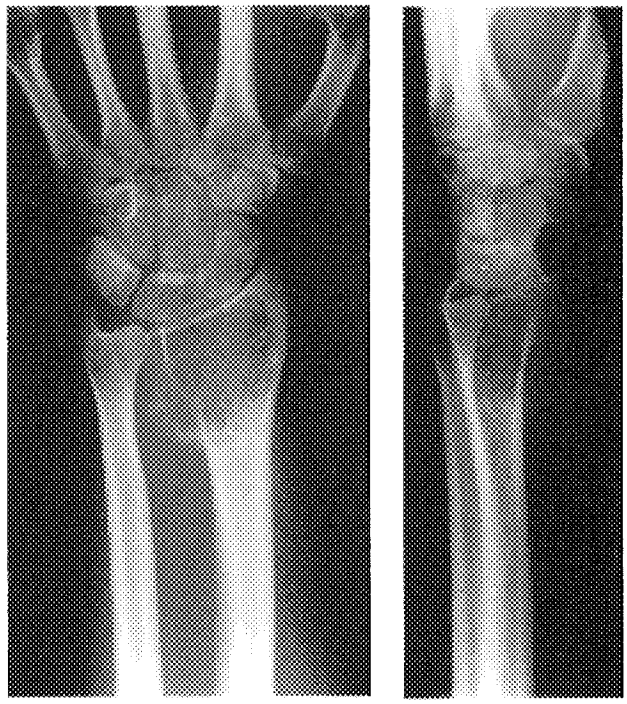

Fig. 6 AP and lateral radiographs showing destruction of the distal radius by a giant cell tumor. The lesion has extended into the soft tissues. This is an aggressive (stage-3) tumor.

(Fig. 6). This staging system is quite useful because it generally dictates the type of treatment necessary for a benign bone tumor. Stage-1 lesions generally are not treated. Stage-2 lesions can be treated successfully with intralesional excision. Finally, stage-3 lesions can be treated with intralesional excision combined with an adjuvant (discussed later) or with marginal or wide en bloc excision.

General Concepts of Treatment

The management of benign bone tumors needs to be individualized on the basis of the special features of each lesion, and it is important to use judgment and experience in determining the best method of treatment. However, certain general concepts apply to the treatment of benign bone tumors. The surgical procedures typically used for the treatment of benign lesions include intralesional excision, marginal en bloc excision, and wide en bloc excision (Table 2).[1] Intralesional excision is defined as a surgical procedure involving entry into the tumor cavity. The technique of intralesional excision, however, has been modernized and differs greatly from the old concept of curettage. This latter procedure often was done through a limited cortical window and was associated with a high risk of local recurrence of the tumor. Currently, intralesional excision means complete exteriorization of the tumor so that there can be complete visualization of the cavity.[3] This usually amounts to unroofing the tumor,

Table 2
Treatment of benign bone tumors

Stage	Treatment
Stage 1	Observation Excision
Stage 2	Intralesional excision with or without adjuvant
Stage 3	Intralesional excision with or without adjuvant Marginal or wide en bloc excision

especially removing all thin cortical areas. The excision is performed with instruments ranging sequentially from large to small. After the lesion has been excised, the excision can be extended with a high-speed burr or with use of adjuvants. The adjuvants available for intraoperative use are liquid nitrogen, phenol, and methylmethacrylate. When these chemicals are placed within the tumor cavity, tissue necrosis occurs to a varying degree beyond the cavity. Liquid nitrogen causes the greatest amount of necrosis; use of this adjuvant can result in necrosis that extends centimeters beyond the cavity wall. This degree of necrosis, however, may lead to fracture after cauterization with liquid nitrogen. Phenol is a cytotoxic substance that is effective in cauterizing the surface of the cavity. The kill is measured in cell layers. Methylmethacrylate can be used both as an adjuvant and as an osseous replacement. As an adjuvant, it kills cells by the heat of polymerization or the toxic effect of the unpolymerized monomer. We believe that the best way to extend the margin of an intralesional excision is with a combination of phenol and methylmethacrylate. In a large multi-institutional study of giant cell tumors treated with intralesional excision, this combination was associated with the lowest prevalence of recurrence.[9]

Marginal and wide en bloc excision are surgical procedures that remove the tumor in one piece. A marginal excision passes through the reactive zone surrounding the tumor, and a wide excision includes a cuff of normal tissue completely encircling the tumor. These types of surgical procedures usually necessitate extensive reconstruction, especially when a joint surface is removed. Reconstruction of an expendable bone, however, is not necessary. These more extensive surgical procedures usually are reserved for aggressive (stage-3) benign tumors or recurrent tumors.

The following 11 benign tumors or tumor-like conditions will be discussed individually, with a focus on their treatment: (1) osteoid osteoma, (2) osteoblastoma, (3) osteochondroma, (4) chondroma, (5) nonossifying fibroma, (6) giant cell tumor, (7) chondroblastoma, (8) fibrous dysplasia, (9) aneurysmal bone cyst, (10) unicameral bone cyst, and (11) eosinophilic granuloma and histiocytosis X. These lesions have been selected because they represent those most commonly seen or because they have special features that need to be considered.

Benign Bone Tumors

Osteoid Osteoma

Osteoid osteoma is a totally benign distinctive lesion, characterized by a richly innervated nidus that is less than 2 cm in diameter and that consists of primitive woven bone and osteoid; the lesion evokes considerable pain and dramatic reactive sclerosis of the surrounding bone. According to Schajowicz,[3] osteoid osteoma accounts for approximately 11% (262 of 2,421) of benign bone tumors and approximately 5% (262 of 5,274) of all primary bone tumors. Most patients who have osteoid osteoma are children or young adults: 70% are less than 20 years old. Osteoid osteoma is rarely found in patients less than 5 years old or more than 40 years old. The lesion was approximately two times more prevalent in boys and men than in girls and women in 262 cases reviewed by Schajowicz.

Diagnosis The pain associated with osteoid osteoma is characterized as being worse at night than during the day. There is evidence that the pain is mediated by prostaglandins and thus may be relieved by aspirin or other nonsteroidal prostaglandin inhibitors.[10] Accompanying abnormalities may include muscle atrophy, a limp, painful scoliosis, or synovitis. Abnormalities of bone growth may occur if the lesion is located near an epiphyseal area.[11] In the course of time, symptoms may subside.[12] Osteoid osteoma occasionally may be asymptomatic, especially when located in the hand.

The most obvious radiographic manifestation is a dense, sclerotic area in a paracortical position, with a central lytic nidus. The actual tumorous tissue is the nidus, so the nidus must be identified for the lesion to qualify for the diagnosis of osteoid osteoma. Plain radiographs give the most information regarding diagnosis.

There are four diagnostic features of osteoid osteoma. The lesion is sharply round or ovoid and usually is less than 1 cm in diameter. The center of the mass is homogeneously dense, and there is a thin, 1- to 2-mm peripheral radiolucent zone. In one study, approximately 50% of 448 osteoid osteomas displayed all of these features.[6] The bones most commonly involved were the femur (135 of 448, 30%), the tibia (121 of 448, 27%), the humerus (45 of 448, 10%), the spine (31 of 448, 7%), and the talus (18 of 448, 4%). In the long bones, the lesion usually is intracortical; in the spine, it usually involves the posterior elements (Fig. 7). Osteoid osteoma of the talus is found more commonly in the neck than in the body.

Osteoid osteoma characteristically has a well-demarcated oval region of bone production separated from the surrounding reactive host woven or lamellar bone. There is no permeation of the lesion into normal bone. The nidus itself contains osteoid or woven bone, or both. There is no evidence of cartilaginous tissue. The center of the nidus usually is more mature and mineralized and therefore has a radiopaque appearance on radiographs.

The differential diagnosis includes bone islands, osteomyelitis, eosinophilic granuloma, osteoblastoma, and osteosarcoma. Differentiation from these entities usually is clear on the basis of the clinical course, radiographic findings, and histologic analysis.

Treatment The standard treatment of osteoid osteoma is complete resection, which immediately relieves the

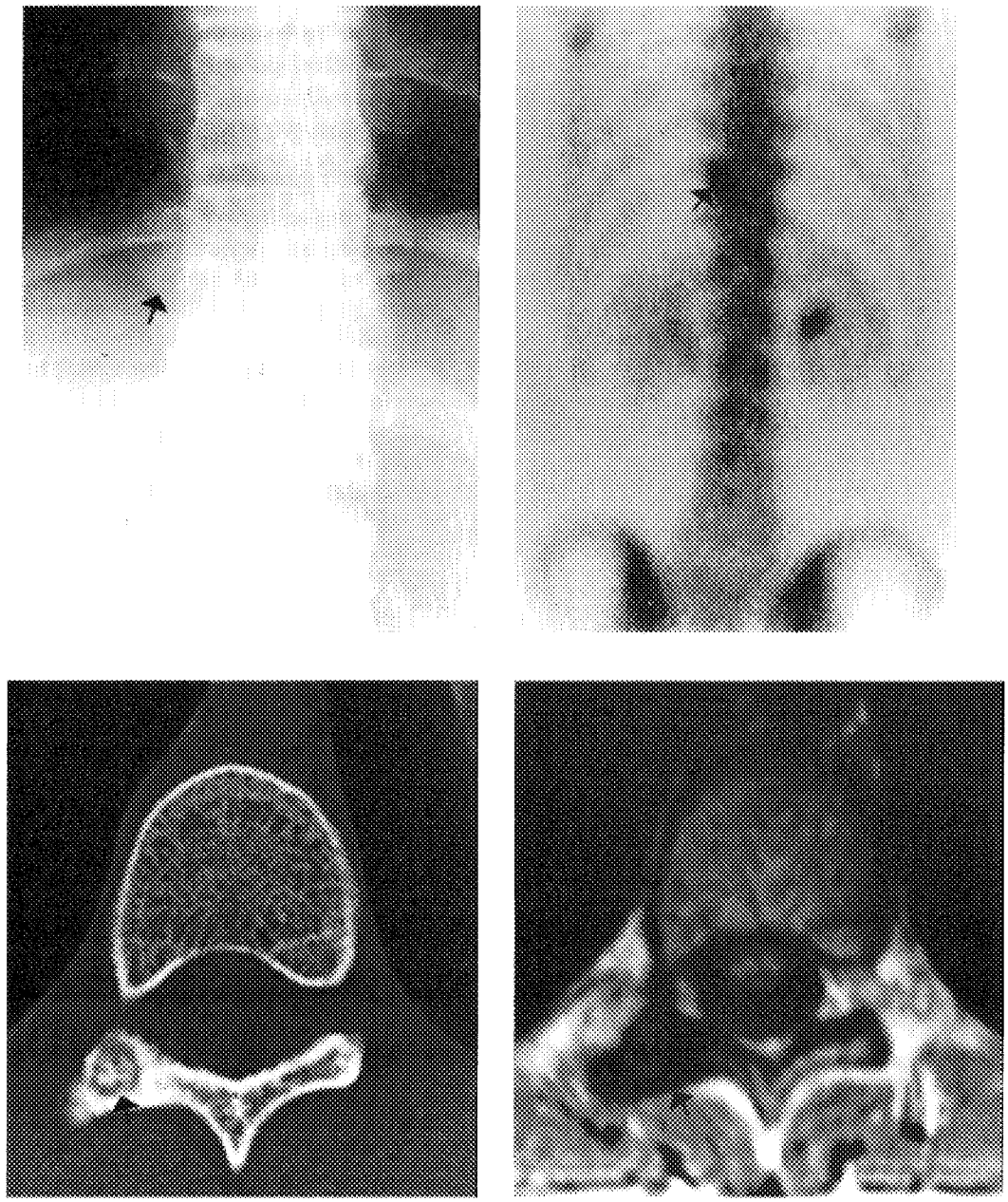

Fig. 7 Top left, AP radiograph of the thoracic spine of a 14-year-old girl who had back pain in the thoracic region. The pain was worse with activity and woke her at night; it was relieved with anti-inflammatory medications. The posterior elements at the tenth thoracic level are enlarged (arrow). **Top right,** Technetium bone scan showing increased activity (arrow) in the area of the tenth thoracic vertebra. **Bottom left,** Thin-section CT scan through the tenth thoracic vertebra, showing a central nidus (arrow) surrounded by sclerotic bone. **Bottom right,** T1-weighted MRI made with contrast medium, confirming the presence of osteoid osteoma (arrow) in the posterior element of the tenth thoracic vertebra. The patient had simple resection without bone grafting or instrumentation. She became asymptomatic and resumed all of her normal activities.

symptoms. Effective resection depends on accurate and specific localization, and many techniques have been used to localize these sometimes evasive lesions. Conventional tomography, nuclear bone-scanning, CT, and gadolinium-enhanced MRI all have been helpful in preoperative localization (Fig. 7). The best method of preoperative imaging appears to be CT. Intraoperatively, tetracycline fluorescence under ultraviolet light may assist in localization as well. Intraoperative nuclear scanning is a reliable technique that can be used to confirm complete removal of the nidus.[13]

Once the nidus has been localized, surgical resection can be planned. The standard method of resection involves removal not only of the nidus but also of a fair amount of surrounding bone in order to be sure that the entire nidus has been resected. While this technique is

effective, it may weaken the bone, necessitate bone grafting, and be complicated by delayed healing and fracture through the site of resection. Several new techniques (described later), such as the burr-down[14] and percutaneous techniques, have been developed to increase the effectiveness of resection and to limit morbidity.

The majority of osteoid osteomas have a palpable, smooth prominence in the area of the reactive bone. The burr-down technique can be used to localize and remove the nidus. Under the guidance of CT scans and intraoperative radiographs, the surgeon makes the operative exposure directly over the suspected site of the lesion. By successively sweeping across the prominent bone with a high-speed burr, the surgeon removes the reactive zone until the hypervascular nidus is encountered. The nidus tissue then is scooped out with a curet and is sent for later histologic examination. The cavity then is enlarged an additional 1 to 2 mm in all directions with the burr. As the deep reactive bone around the nidus is, for the most part, preserved, there is minimum weakening in the area. Complications and recurrence are minimum with this method.[14]

Percutaneous procedures also have been used to treat osteoid osteoma. With the use of drills guided by CT, osteoid osteomas may be removed in the radiology suite. While this method appears to be effective clinically, it does not reliably retrieve diagnostic tissue for analysis.[15] Another technique uses percutaneous radiofrequency electrodes to ablate areas of presumptive osteoid osteoma. This technique has been used on a limited basis.[16] Again, with this technique, tissue is unavailable for histologic analysis. In addition to the surgical treatment of osteoid osteoma, a medical regimen involving prostaglandin inhibitors has been proposed.[12] While this approach makes sense empirically, the treatment protocol is prolonged and has had variable results in our hands. As the burr-down technique has minimum morbidity and excellent results with quick recovery, it is our treatment of choice for easily accessible lesions.

Osteoblastoma

Osteoblastoma is a relatively rare benign lesion that is larger than 2 cm in diameter and is well confined within the bone. The lesion is composed mostly of osteoid and woven bone. It has been called a giant osteoid osteoma.[3]

Schajowicz[3] reported that osteoblastoma accounts for approximately 3% (69 of 2,421) of benign bone tumors and 1% (69 of 5,274) of all primary bone tumors. The peak prevalence of osteoblastoma is in patients between 20 and 30 years old, and in Schajowicz's study 62 (90%) of the 69 patients who had osteoblastoma were less than 40 years old. The age distribution is similar to that of osteosarcoma.[11]

Diagnosis Pain is the predominant symptom of osteoblastoma. The symptoms are present for a relatively long time, and the pain is characterized less easily than that associated with osteoid osteoma. If the lesion is located in the spine, it may cause scoliosis or neurologic symptoms, or both. Osteoblastoma normally centers in the medullary portion of bone, and the growth of the tumor usually results in fusiform expansion. A thin rim of periosteal new bone usually surrounds the lesion. The center area may be lucent or blastic, or it may have a mixed appearance. The most common site of osteoblastoma is the spine, particularly the posterior elements; Mirra and associates[6] reported that 60 (42%) of 144 such lesions were found in the spine. Other common locations are the long bones of the extremities.

Osteoblastoma is characterized by the production of pure osteoid and woven bone. No production of lamellar bone is seen. There is no evidence of permeation into host lamellar bone. Benign-appearing osteoblasts are found in close association with the forming osteoid and woven bone. This osteoblastic rimming is characteristic of osteoblastoma and is seen rarely in high-grade osteosarcoma.[6] There is no production of cartilaginous tissue.

The differential diagnosis includes osteoid osteoma, osteosarcoma, giant cell tumor, and aneurysmal bone cyst. Careful pathologic assessment of the tissue taken for biopsy should be performed to exclude the rare case of concomitant osteoblastoma and osteosarcoma.[6]

Treatment If osteoblastoma can be excised completely, local control usually is achieved. Complete excision can be accomplished by a marginal or intralesional resection with use of curettage. Portions of bone that can be easily removed and discarded, such as the fibula or rib, are best treated with en bloc resection. It is often difficult to perform an adequate surgical procedure in the spine because of the proximity of neural structures. Adjuvant treatment, such as cryosurgery, radiation, or chemotherapy, may be used in such cases.[17-19]

Osteochondroma

Osteochondroma is a benign developmental growth defect, found in the metaphyseal area of long bones, that produces a lesion with an osseous base and a cartilage cap. Evidence suggests that the lesion is produced by aberrant epiphyseal plate cartilage, as its histologic appearance is similar to that of a growth plate. Osteochondromatosis is a hereditary autosomal dominant condition characterized by multiple osteochondromas involving multiple sites.

Osteochondroma is the most common benign skeletal lesion. It has been reported to account for 44% (1,064 of 2,421) of benign tumors and 20% (1,064 of 5,274) of all primary bone tumors.[3] A slight male predominance has been described (male-to-female ratio, 1.6:1).[4] Osteochondroma is found more frequently in the lower extremity than in the upper extremity, and 36% (355 of 983) of the lesions are distributed around the knee.[3,20] In the report by Schajowicz,[3] 69% (687 of 996) of the

solitary osteochondromas were found in patients who were less than 30 years old. Multiple osteochondromas usually are found in patients who are less than 20 years old.[6]

Osteochondroma may or may not be symptomatic, depending on its location. If it arises in the area of a tendon or a muscle belly, chronic irritation may produce symptoms. Osteochondroma in the vicinity of a joint may limit range of motion. Additional causes of symptoms are nerve compression, fracture of the stalk, or the development of a pseudoaneurysm.[6]

Osteochondroma has characteristic radiographic features. The lesion protrudes from the surface of the bone and may be either sessile or pedunculated. The architecture of the lesion bone blends imperceptibly with that of the host bone. The outer surface of the lesion is rounded or lobulated. The size ranges from small (2 cm or less) to quite large (12 cm or more). The stalk of a pedunculated osteochondroma is composed of thin cortical bone, the medullary cavity of which is contiguous with the medullary cavity of the underlying host bone.

The primary histologic characteristic of osteochondroma is a cartilaginous cap. The cap is of variable thickness; usually it is intact in children and thinned in adults. The cartilage in the cap is benign histologically; occasionally, there is increased cellularity, and double-nucleated chondrocytes may be found.[6] Because this lesion represents abnormal enchondral ossification, it has many features of a typical growth plate.

The differential diagnosis includes parosteal osteosarcoma and malignant cartilaginous exostosis.[6] Histologically, parosteal osteosarcoma is composed of dense fibro-osseous tissues, which are not present in osteochondroma. The cartilage cells in a malignant cartilaginous exostosis show typical changes of malignancy.

Treatment If an osteochondroma is found incidentally and is small, no treatment is necessary. An osteochondroma that is symptomatic because of its position can be treated with simple resection at the base of the osseous abnormality. As long as all of the cartilaginous tissue, including the perichondrium surrounding the cap of the lesion, is removed, there should be no recurrence. A symptomatic osteochondroma of the spine should be resected, and improvement can be expected in most patients.[21]

Malignant Transformation Malignant transformation probably occurs in fewer than 1% of osteochondromas.[6] A lesion that is wide-based or sessile has a considerably greater surface area and thus is more likely to undergo malignant transformation. Patients with multiple osteochondromatosis are at an increased risk for the development of chondrosarcoma because of the numerous lesions. Malignant transformation should be suspected if the lesion suddenly becomes symptomatic, begins to grow rapidly, or has a cartilage cap with a thickness of more than 1 cm or with a maximum diameter of more than 5 cm (Fig. 8).[1] Traditionally, CT has been used for imaging of suspicious osteochondromas. However, this modality is unreliable in estimating the thickness of the cartilaginous cap.[20] MRI is much more accurate in measuring the cartilaginous cap and in identifying the presence of a stalk by showing marrow that is continuous between the host bone and the lesion. If there is any question regarding the possible malignant transformation of an osteochondroma, the lesion should be resected.

Chondroma

Chondroma is a lesion of mature hyaline cartilage that may be one of two types: (1) enchondroma, which is located centrally within the bone, or (2) periosteal or cortical chondroma, which arises either in or beneath the periosteum. The lesion is characterized by mature, benign cartilage tissue. Malignant transformation occurs rarely.

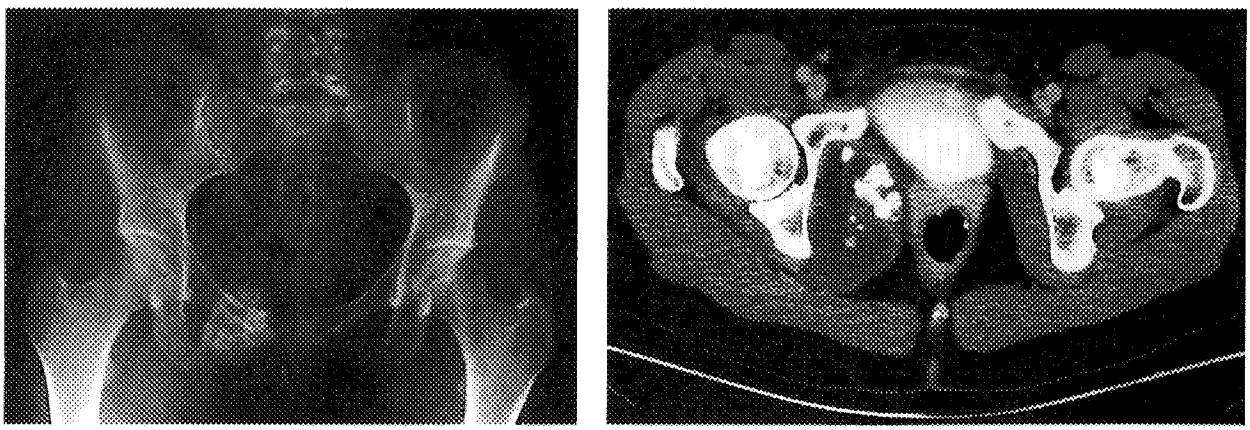

Fig. 8 **Left,** AP radiograph of the pelvis of a 24-year-old woman with pain in the right side of the groin, showing a lesion that was thought to be a benign osteochondroma. **Right,** Subsequent CT scan shows a large soft-tissue mass in the pelvis that was consistent with chondrosarcoma.

Chondroma has been reported to account for 25% (616 of 2,421) of benign bone tumors and 12% (616 of 5,274) of biopsy-proven primary bone tumors.[3] Chondroma is detected throughout life. Patients with multiple lesions usually are between 10 and 30 years old, with the peak prevalence occurring during the third decade of life. There is no sex predilection for skeletal chondroma.

Enchondroma is quite common in the small bones of the hands and feet, with 58% (89 of 153) of the lesions in one study occurring in these areas.[6] Indeed, enchondroma is the most common primary tumor found in the bones of the hand. Asymptomatic enchondromas usually are found incidentally on radiographs. Atypical enchondromas are characterized as such because of their association with a history of pain.[6] A fracture sometimes is seen in the area of enchondroma, perhaps because of previous weakening of the bone or simply as an incidental finding.

Occasionally, a patient has multiple enchondromatosis, or Ollier disease. A patient who has multiple enchondromas and angiomas of the soft tissues has Maffucci syndrome. Sarcomatous transformation of the enchondromas can occur in association with both of these syndromes.

Periosteal chondroma is a rare, benign cartilaginous tumor that arises from the periosteal tissues. It is the periosteal counterpart of medullary enchondroma. Additional names for this lesion are parosteal chondroma and juxtacortical chondroma.[6] The signs and symptoms of periosteal chondroma are similar to those of solitary enchondroma. Pain is the most common symptom, and the lesion usually is found on an incidental basis. It rarely, if ever, converts to a malignant lesion.

Enchondroma Classically, enchondroma is a long, oval lesion that is located centrally in the tubular portion of the bone. It usually has a lobulated appearance grossly and typically is sharply demarcated. Calcification of the

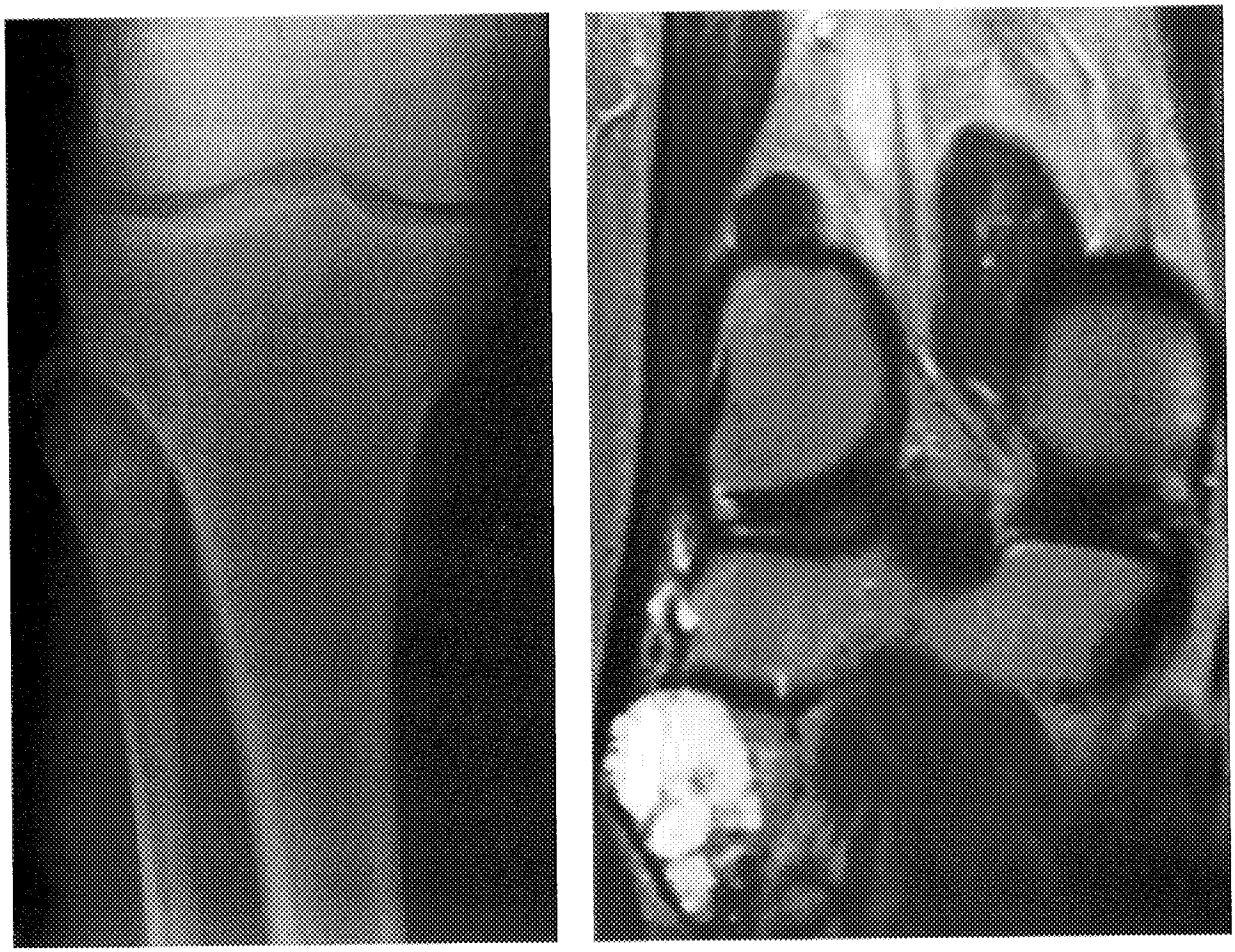

Fig. 9 AP radiograph and sagittal T2-weighted MRI of a 30-year-old woman with pain in the lateral aspect of the knee. There is a destructive-appearing cartilaginous lesion in the proximal fibula. Because of the invasive nature of the lesion, an en bloc resection was performed without biopsy.

lesion may or may not be present. Expansion of the surrounding cortex is uncommon unless the lesion occurs in a small bone, such as the bones of the hands or feet, or in the fibula.

Malignant transformation of enchondroma is rare; according to Mirra and associates,[6] who did not give actual numbers, malignant transformation occurs in fewer than 1% of these lesions. Radiographic features of malignant transformation include increased biologic activity, as evidenced by increased endosteal scalloping, pathological fracture, or increased size. Bone-scanning of the area may or may not indicate increased activity and is not a reliable way to assess malignant transformation. MRI occasionally is helpful in assessing the nonmineralized component of the lesion and its apparent aggressiveness.[20] If the imaging studies and clinical situation indicate the potential for malignant transformation, biopsy is mandatory (Fig. 9).

The differential diagnosis includes epidermoid cyst in the distal phalanx and fibrous dysplasia, nonossifying fibroma, simple bone cyst, chondroblastoma, chondrosarcoma, chondromyxoid fibroma, and bone infarction elsewhere. Usually, these lesions can be excluded on the basis of radiographic examination. If not, biopsy may be necessary.

Enchondromas are composed of benign cartilaginous cells. Signs of possible malignant transformation, such as increased cellularity and occasional binucleated cells, are compatible with a benign diagnosis, especially in the hands and feet, but the differentiation between enchondroma and grade-1 chondrosarcoma can be quite difficult histologically. The cytologic features of enchondroma and grade-1 chondrosarcoma often overlap.[6] The size of the nuclei, the number of cells per unit of area, the number of double-nucleated chondrocytes, and the number of mitoses all are important characteristics that must be evaluated. The distinction between enchondroma and low-grade chondrosarcoma is not made strictly on the basis of histologic criteria; radiographic criteria are used as well. Intimate communication should take place between the surgeon, the pathologist, and the radiologist to formulate the final diagnosis.

Periosteal Chondroma The features of periosteal chondroma include an eccentric, juxtacortical, longitudinally oriented mass that is 1 to 3 cm in size and that contains small rounded densities characteristic of cartilage (Fig. 10). The cortical border shows a rind of osseous sclerosis.[6] If these features are not evident, the possibility of a malignant tumor must be considered. Two-thirds of these lesions occur in the center of the metaphysis, and the remaining one-third arise in the diaphysis.[3,6] By histologic definition, periosteal chondroma must produce hyaline cartilage. On occasion, myxoid changes may be found. There is no invasion into adjacent tissue or nuclear anaplasia.

Treatment of Enchondroma An atypical or symptomatic enchondroma should be treated locally with marginal or intralesional resection. The tissue should be analyzed quite carefully for any histologic changes suggestive of malignancy. Defects created in large and weightbearing bones should be grafted with either autologous or allograft material.[22,23] Bone grafting may not be necessary after curettage of an enchondroma of the hand.[24]

Treatment of Periosteal Chondroma Periosteal chondroma is a benign lesion. Marginal or wide local resection is the procedure of choice. However, if the lesion is excised incompletely, it may recur. Bone grafting may or may not be necessary, depending on the extent of the local resection.

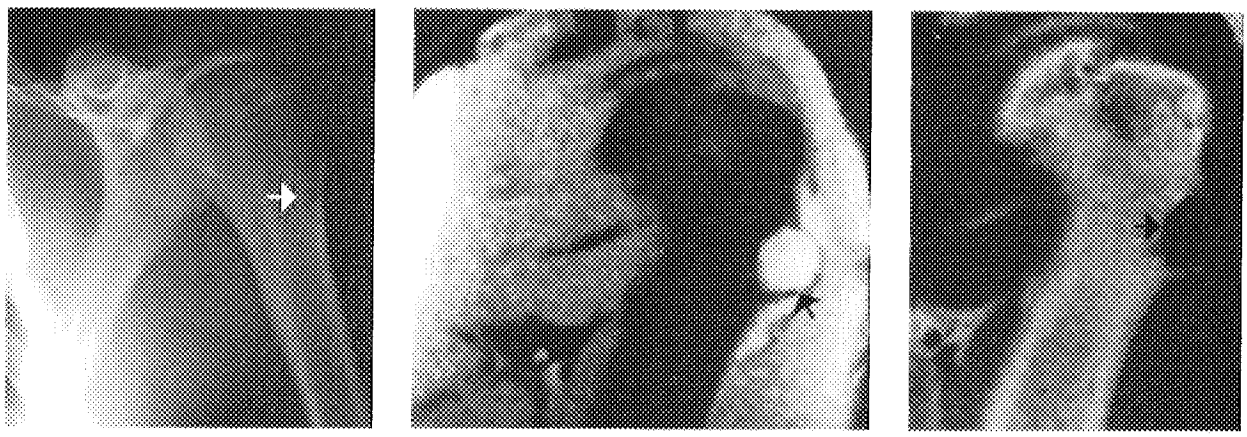

Fig. 10 Left, AP radiograph of a 22-year-old man with pain in the shoulder shows a surface lesion (arrow) of the proximal part of the humerus laterally. **Center** and **right,** The lesion (arrows) as it appears on coronal T2-weighted and T1-weighted MRIs of the involved area. These studies were interpreted as showing periosteal chondroma. After confirmation on frozen section of a benign cartilaginous lesion, simple resection was performed and was curative.

Nonossifying Fibroma and Fibrous Cortical Defect

Nonossifying fibroma and fibrous cortical defect are histologically identical; they are benign tumors with slightly different characteristics. Fibrous cortical defect is asymptomatic and usually is solitary, with a predilection for the diaphysis of long bones. It is quite common—Mirra and associates[6] estimated that it is found in 30% to 40% of all children between 4 and 8 years old—and it rarely needs treatment. The etiology is unknown. Nonossifying fibroma also usually is a solitary lesion, although patients occasionally have multiple areas of involvement. Most of these lesions do not cause symptoms. Unlike fibrous cortical defect, nonossifying fibroma is found most often in the metaphyseal area (Fig. 11, *left*). It only becomes symptomatic if it is large enough to be a major stress-raiser in the bone or to cause a pathologic fracture.

Most lesions are discovered incidentally on radiographs made for other purposes. Eighty percent of nonossifying fibromas are discovered in patients who are less than 20 years old. The male-to-female ratio is approximately 1.4:1 (88:62).[6] Nonossifying fibroma accounts for approximately 2% of primary bone tumors undergoing biopsy.[3]

Most of these lesions do not cause symptoms. There are three phases in their growth: early, middle, and regressive. Lesions in the early and middle phases are found in patients less than 20 years old. Those lesions usually are eccentric and cause symptoms only if the bone is weakened sufficiently for microfractures or stress fractures to occur in the area. After skeletal maturity, these tumors go into the regressive phase and heal; they are seen very rarely in patients who are more than 35 years old.[6]

Nonossifying fibroma usually is eccentric in its location; this lesion most often involves the medullary canal and the overlying cortex. It is located in the metaphyseal or the metadiaphyseal area and exhibits a longitudinal growth pattern. The outer edges of the lesion are lobulated. The internal architecture has a bubbly appearance. There usually is a rind of reactive bone or sclerosis surrounding the lesion; however, there may be cortical thinning, which may be misinterpreted as a malignant change.

Diagnosis A bone scan usually shows minimally to mildly increased uptake, which may be very difficult to distinguish because of the adjacent growth plate. Generally, plain radiographs are diagnostic and additional imaging is not necessary. A CT scan, however, may show additional cross-sectional anatomy and assist in the diagnosis. MRIs, when done, generally show a low signal on both T1 and T2-weighted studies; this reflects the fibrous-tissue content of the lesion.[25]

Nonossifying fibroma usually is brown, yellow, or grayish and is composed of soft or firm fibrous tissue. Histologically, the lesion comprises fibrous tissue, xanthoma cells, and multiple giant cells. In the early phase, a spindle-cell stroma also may be in evidence. As

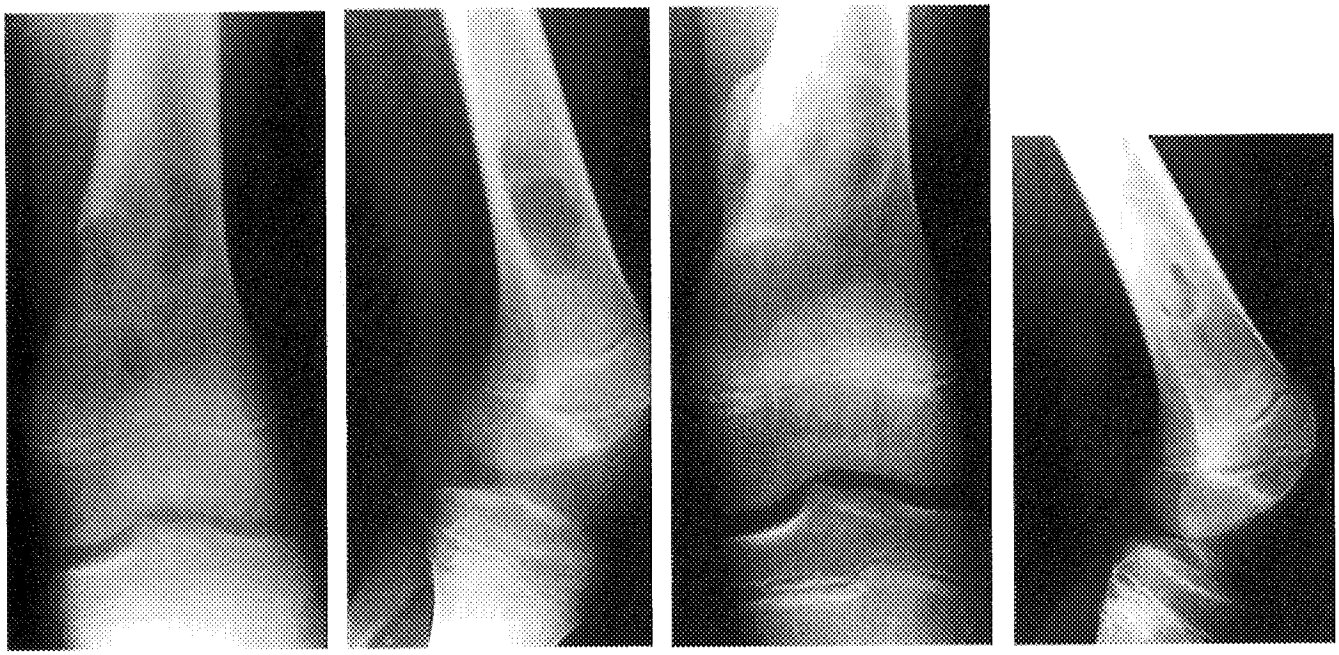

Fig. 11 Left, AP and lateral radiographs of a 13-year-old boy with pain in the distal femur were consistent with a large nonossifying fibroma at risk for pathologic fracture. **Right,** AP and lateral radiographs made 6 months after curettage and grafting with demineralized bone matrix.

the lesion matures through the middle and regressive phases, there is increased production of collagen, a decrease in the number of giant cells, and an increase in the number of lipid-rich macrophages.[6] Hemosiderin pigment is in evidence throughout the life of the lesion but increases with maturation. The differential diagnosis of these tumors includes brown tumor of hyperparathyroidism, Paget disease, fibrous histiocytoma, desmoplastic fibroma of bone, osteosarcoma, and osteofibrous dysplasia.

Treatment Asymptomatic lesions need not be treated. A very large lesion theoretically may weaken the bone and may need to be treated prophylactically. It has been proposed that a nonossifying fibroma that involves more than 50% of the width of the bone on plain radiographs in two projections or that is more than 33 mm in length may cause fracture and should be treated.[26] If the lesion requires treatment, local intralesional curettage with bone grafting is sufficient (Fig. 11, *right*). To obviate the morbidity associated with autologous grafting, allograft tissue may be used.[23,27] If a pathologic fracture occurs through a nonossifying fibroma, the fracture may promote healing and may lead to more rapid regression of the lesion, thereby averting the need for surgery.

Giant Cell Tumor of Bone

Benign giant cell tumor is a low-grade neoplastic lesion that almost always arises in the epiphyseal or metaphyseal region of a long bone and develops through the process of enchondral ossification. The lesion usually is solitary and becomes symptomatic when enough bone stock has been destroyed.

Benign giant cell tumor accounted for approximately 19% (460 of 2,421) of benign bone tumors and 9% (460 of 5,274) of all primary bone tumors in the review by Schajowicz.[3] He also reported that 75% (345 of 460) of patients who have a giant cell tumor are 20 to 50 years old. Most giant cell tumors occur after the epiphyseal plate has closed. Thus, giant cell tumors arise more often in girls who are less than 17 years old than in boys of the same age. Overall, there is a slight female-to-male preponderance.[6] There is an increased prevalence of giant cell tumor of the spine, particularly in girls and women, in the second and third decades.[28] By far, most giant cell tumors occur about the knee, in the proximal humerus, and in the distal radius.

Diagnosis Pain without an obvious pathologic fracture has been the most frequent presenting symptom, in our experience. In the remainder of patients, a pathologic fracture has been the presenting complaint. Local pain, swelling, and tenderness are seen in most patients. Serum calcium levels are normal, not elevated as they often are with brown tumors of hyperparathyroidism. Systemic complaints are uncommon. Patients with involvement of the spine or sacrum may have neurologic signs and symptoms.

Radiographs usually reveal the lesion to be quite destructive of both medullary and cortical bone (Fig. 12). The lesion is characterized by an expanding zone of radiolucency that is located eccentrically in the end of a long bone in an adult.

When a giant cell tumor escapes the boundaries of the cortex, a soft-tissue mass may be present. The lesion may extend to and involve the subchondral bone in a periarticular arc. Although CT scanning is quite helpful in detecting thinning of the bone and evaluating an associated thin rim of bone surrounding the lesion, it is not as effective as MRI in evaluating subchondral cortical penetration, joint involvement, and pathologic fracture.[29] MRI is the most useful technique, morphologically, for determining the extent and stage of a local giant cell tumor. Bone-scanning is useful for detecting multicentric giant cell tumor, which is rare. (Less than 1% [4 of 460] of the giant cell tumors in the study by Schajowicz[3] were of this type.)

Location is very important in the diagnosis of benign giant cell tumor. Although giant cell tumor historically has been described as an epiphyseal tumor, more recent evidence has indicated that its actual origin may be on the metaphyseal side of the healed epiphysis.[30]

The color of the tissue removed from a benign giant cell tumor generally ranges from brownish tan to yellow. Areas of hemorrhage may be dark red. Areas of necrosis may appear cyst-like. Histologically, a benign giant cell tumor contains predominantly osteoclast-like giant cells and spindle-shaped stromal cells. The nuclei in both of these cell types are round or oval and are uniform in size, with blandly granular chromatin and prominent nucleoli.[6] The giant cells never show mitotic figures; however, the stromal cells can show five to ten mitoses per ten high-power fields. It is not possible to predict the biologic behavior of a particular tumor on the basis of its histologic appearance.

Giant cell tumors exhibit a wide variety of biologic activity. Some lesions act quite benign, remain local and noninvasive, and do not metastasize; others are extremely destructive locally or metastasize to the lungs. More sophisticated studies, such as flow cytometric analysis of DNA, have not been helpful in predicting whether a tumor will metastasize.[31]

In an effort to better predict the biologic activities of tumors, Campanacci and associates[32] developed a staging system, later modified,[33] that was based on a combination of clinical, radiographic, and pathologic findings. A stage-I giant cell tumor is defined as one that causes symptoms, appears latent radiographically, and has a benign histologic pattern. A stage-II giant cell tumor may cause symptoms, has an active radiographic appearance without evidence of metastasis, and has a benign histologic pattern. A stage-III giant cell tumor causes symptoms, has radiographic signs of rapid and invasive growth with extracortical and subchondral extension, and often

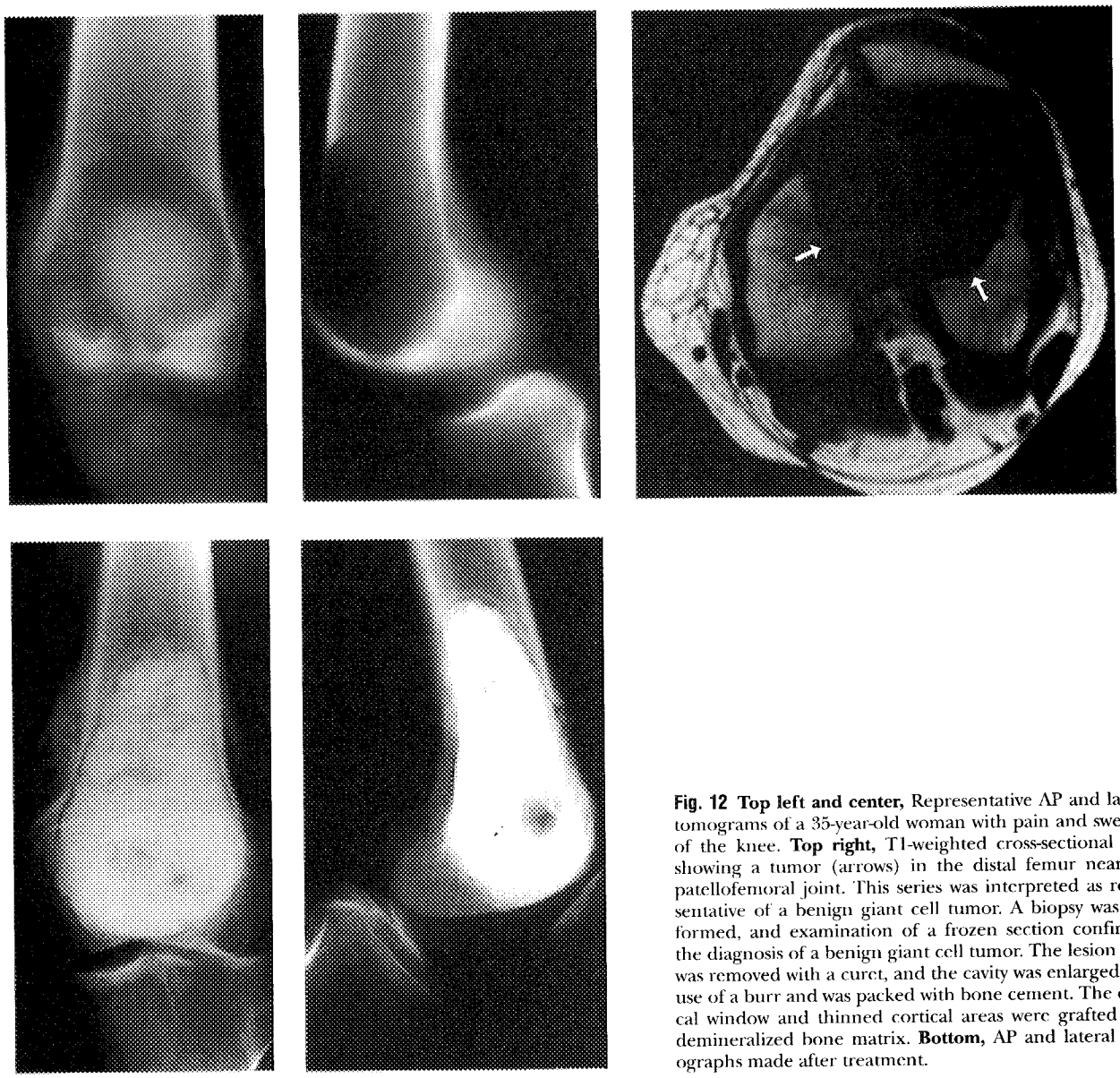

Fig. 12 Top left and center, Representative AP and lateral tomograms of a 35-year-old woman with pain and swelling of the knee. **Top right,** T1-weighted cross-sectional MRI showing a tumor (arrows) in the distal femur near the patellofemoral joint. This series was interpreted as representative of a benign giant cell tumor. A biopsy was performed, and examination of a frozen section confirmed the diagnosis of a benign giant cell tumor. The lesion then was removed with a curet, and the cavity was enlarged with use of a burr and was packed with bone cement. The cortical window and thinned cortical areas were grafted with demineralized bone matrix. **Bottom,** AP and lateral radiographs made after treatment.

is accompanied by a soft-tissue mass. The histologic findings in stage III are still benign. Several studies have showed this system to be predictive; however, it has not been helpful in planning treatment, as some stage-III lesions can be treated adequately with local curettage while others require en bloc resection.

The differential diagnosis of giant cell tumor includes brown tumor of hyperparathyroidism, malignant fibrous histiocytoma, nonossifying fibroma, chondroblastoma, osteoblastoma, and osteosarcoma. Careful inspection of the complete specimen is mandatory to rule out areas of osteosarcoma.

Treatment The treatment of giant cell tumor is controversial, and the literature is confusing. Traditionally, giant cell tumor has been treated with curettage or intralesional resection, which has resulted in a very high rate of recurrence with a range of 35% (77 of 218)[34] to 42% (22 of 53).[35] Because these figures suggest the inadequacy of the local procedure, several surgeons have turned to a more extensive approach involving en bloc resection, reducing the rate of local recurrence to approximately 10%.[6] En bloc resection is a major procedure that usually involves the use of massive allografts (Fig. 13). While the rate of recurrence associated with this technique is quite low, complications such as infection, resorption, collapse, and fracture cumulatively produce a high rate of complications (13 [24%] of 55 in one study[36] and 13 [65%] of 20 in another[37]). One must

weigh the risk of recurrence following curettage against the risk of complications and a poor functional result associated with en bloc resection. The problem with simple curettage is that a giant cell tumor forms contiguous pockets of tumor in both medullary and cortical bone; this creates an irregular, ill-defined margin. Simple curettage usually leaves residual microscopic evidence of disease. Several authors have reported using local extension modalities (phenol, liquid nitrogen, and carbon-dioxide lasers) to kill these residual microscopic foci.[38-41] Although it makes some sense to try to extend the margin locally with these various modalities, the actual effect of phenol and carbon-dioxide lasers is unclear. Cauterization with liquid nitrogen does extend the margin by circumferential necrosis; however, the complications of burns of the skin and delayed pathologic fracture occur quite frequently with this technique.[40]

The basic goal in the local treatment of giant cell tumor is to remove adequate tissue without destabilizing the area or creating the need for a major reconstruction. Over the last decade, a technique combining more extensive local excision with use of polymethylmethacrylate cement has become a popular way to achieve this goal.[9] The modern technique for the removal of a giant cell tumor involves wide decortication, or windowing, of all bone overlying the area of the tumor (Fig. 14, *A* and *B*). This allows direct visualization of the whole tumor cavity (Fig. 14, *C*). Conventional curets as well as a high-speed burr are used to remove medullary and cortical bone. When the resection has been completed, nothing should be visible except normal cortical and medullary bone. Extensive irrigation also should be used. Because the cavity will be filled with methylmethacrylate bone cement to achieve immediate stability (Fig. 14, *D* and *E*), the surgeon is much more likely to remove additional tissue to ensure complete resection. Initially, it was thought that the heat of polymerization of the methylmethacrylate extended the margins of the local resection. However, this remains controversial.[42,43] In fact, the heat that is generated may cause necrosis of the immediately adjacent subchondral bone and articular cartilage; therefore, the adjacent articular surfaces should be irrigated with cooled saline solution as the cement is hardening to prevent heat necrosis of the articular cartilage. After the cement has hardened, the cortical window is grafted (Fig. 14, *F*) or covered with demineralized bone matrix to stimulate the restoration of strong cortical boundaries. Unless it creates a problem later, the cement is not removed.[43]

In patients in whom a major pathologic fracture has occurred or there is invasion of the articular structures, en bloc resection and use of an osteochondral allograft should be considered, especially when the articular surfaces cannot be reconstructed using the cementation technique. Usually, only a portion of the articular surface needs to be reconstructed. The resection should consist of a marginal or wide local resection, with removal of all involved tissues. The reconstruction of the defect with large frozen allografts is technically difficult. Attention should be paid to creating exact cuts in the bone and to achieving good apposition of host and donor bone as well as rigid internal fixation. Bone

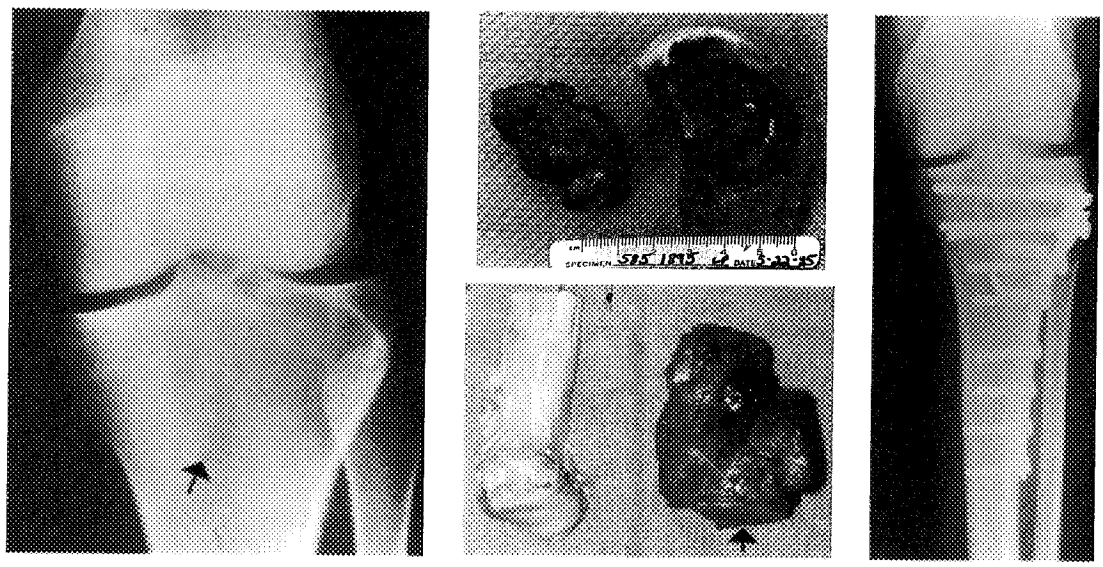

Fig. 13 **Left,** AP radiograph of a 30-year-old woman with a biopsy-proven benign giant-cell tumor (arrow) of the proximal tibia with an intra-articular fracture. **Top center,** The resected lateral tibial condyle. **Bottom center,** Specimen (arrow) with allograft osteoarticular replacement. **Right,** AP radiograph made 5 years postoperatively. The patient had mild crepitus, no pain, and excellent motion and function.

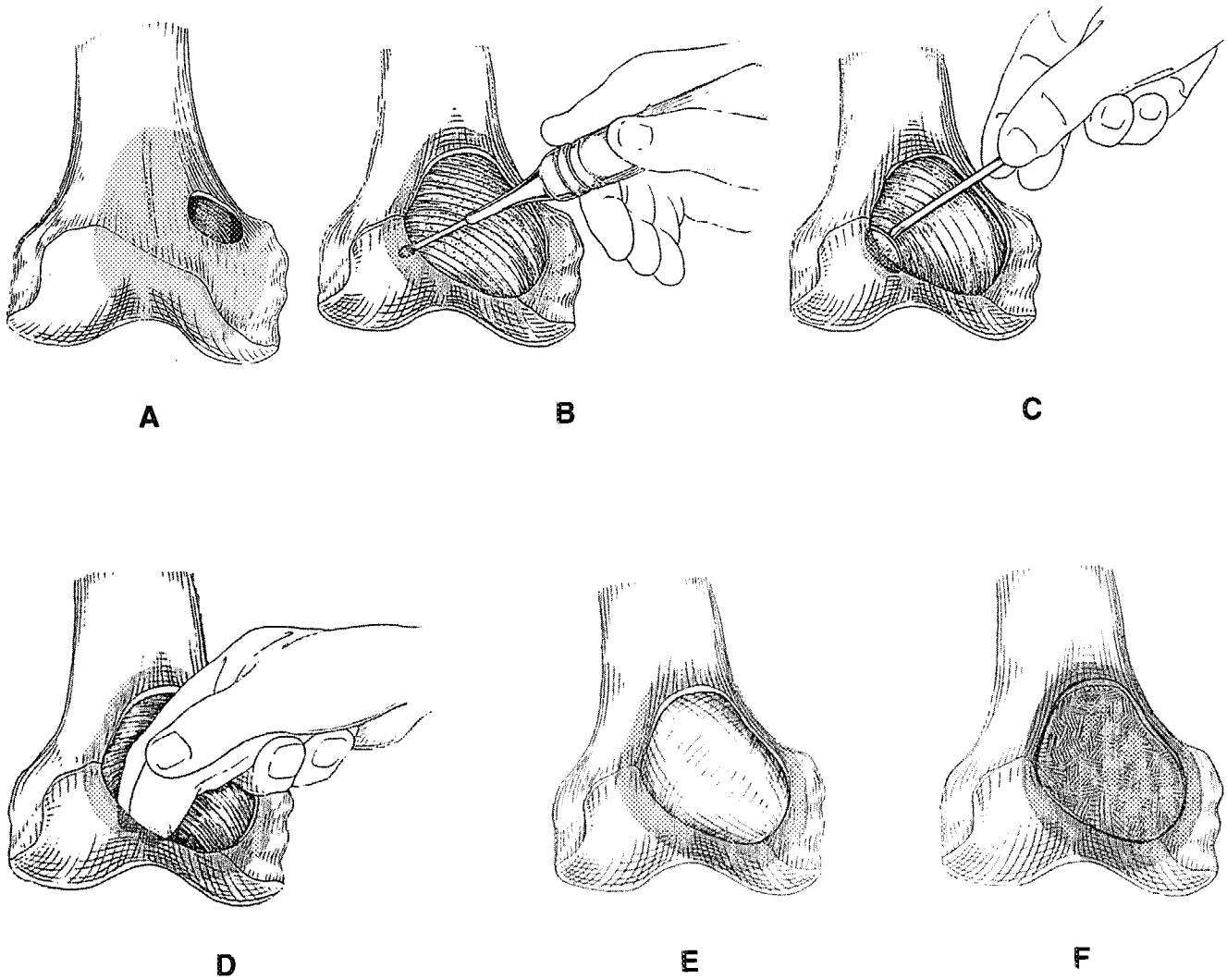

Fig. 14 Treatment of benign giant cell tumor. **A,** Benign giant cell tumor of the distal femur with biopsy defect. **B,** The cortical opening is enlarged to allow curettage and high-speed burring to normal bone under direct vision. **C,** A dental mirror is used to ensure margins of normal bone throughout the entire cavity. **D,** Bone cement is digitally packed into the margins of the cavity. Iced saline solution is used to irrigate the area and joint surface to prevent thermal damage to the articular surface. **E,** The cavity is filled with cement and an indentation is made for the subsequent bone graft. **F,** The bone graft is placed on the surface of the cement, filling the cortical defect.

grafting at these sites probably decreases the prevalence of nonunion. Meticulous attention also should be paid to achieving adequate soft-tissue coverage and closure.

Occasionally, giant cell tumor occurs in an area such as the sacrum[44] in which resection is extremely difficult or impossible; such tumors have a very high rate of recurrence. Local and meticulous curettage remains the principal treatment of sacral tumors. Every attempt should be made to preserve neurologic function. If both nerve roots of the second sacral vertebra and one nerve root of the third can be saved, the patient should maintain normal bowel, bladder, and sexual function.[42] Wide local resection involving both an anterior and a posterior approach may result in severe impairment of distal neurologic function and of spinal stability and should be reserved for patients who have an exceptionally large tumor or a tumor in which there is malignant degeneration. Cryotherapy may have a place in the treatment of sacral tumors; even though sacral nerve roots are frozen in the process, they may regenerate.[40]

Most authors have agreed that radiation therapy should be avoided in the treatment of giant cell tumor, as there is a high prevalence of sarcomatous degeneration.[4] While more recent studies have indicated that modern radiation techniques can be used to treat benign giant cell tumor of bone more effectively,[45] these

studies are in an early stage and the follow-up period is too short to evaluate the prevalence of secondary sarcomatous degeneration adequately. Radiation therapy should be reserved for patients in whom complete resection is impossible.

Chondroblastoma

Diagnosis Chondroblastoma typically is an epiphyseal tumor that occurs in adolescents; it occurs slightly more commonly in boys than in girls.[9] Approximately two-thirds of these lesions occur in the proximal humerus, the distal femur, or the proximal tibia. Patients typically have a painful cystic erosion of the epiphysis or metaphysis, and this particular lesion is one of the few benign tumors that has a small risk of associated pulmonary nodules.[4] The pulmonary metastases have the same histologic appearance as the primary lesion. On imaging studies, these epiphyseal abnormalities are lucent, with or without intrinsic, fine calcifications within the lesion itself (Fig. 15).[46] The Enneking[1] stage of chondroblastoma usually is 2 or 3.

Treatment The recommended treatment of chondroblastoma includes a biopsy, to document the histology, followed by curettage. Intralesional curettage is best combined with local adjuvant treatment such as freezing with liquid nitrogen or application of phenol. The defects created by this lesion require a local graft of either autogenous or allogeneic bone. Because of the subchondral erosions typically produced by this tumor, reconstruction of joint surfaces may need to be considered.

Overall, the prognosis is good for most patients; however, the rate of local recurrence was 14% (seven of 50) in one study.[47] This relatively unusual tumor requires careful surgical treatment with the aim of avoiding local recurrence and subsequent loss of joint function. Careful preoperative imaging will improve surgical results. If pulmonary nodules occur, they should be removed whenever possible.

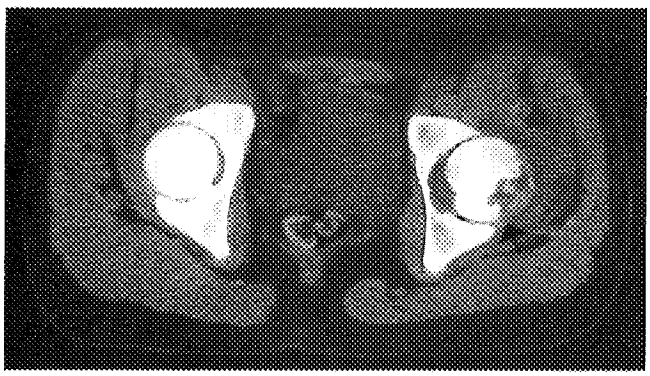

Fig. 15 CT scan of the pelvis of a 14-year-old with left hip pain, showing a lucency of the femoral head (epiphysis). A typical subchondral erosion in chondroblastoma is evident (arrow).

Fibrous Dysplasia

Fibrous dysplasia is a disease process in which normal bone is replaced with dysplastic bone permeated by fibrous tissue.[48] It is diagnosed most commonly in adolescents, although approximately one fourth of the lesions occur initially in adults. In our experience, children who presented at a younger age had a slightly worse prognosis, with more extensive involvement and a higher prevalence of fractures or bone pain. The tumor occurs slightly more often in girls and women than in boys and men and has a polyostotic presentation in one fourth of the patients. An unusual combination consisting of precocious puberty, polyostotic disease, and café-au-lait markings on the skin constitutes the findings of Albright syndrome. The most common sites of monostotic involvement include the proximal femur, the tibia, the humerus, the ribs, the skull, and the cervical spine.[49]

Diagnosis Fibrous dysplasia presents with a myriad of radiographic findings, but most typical and classic is the ground-glass appearance, which results from the fibrous stroma replacing bone (Fig. 16). Fibrous dysplasia also may present as a cystic lesion in the proximal femur, or even as a densely sclerotic lesion of bone. Occasionally, expansion of the involved segment of bone is found, and, particularly in earlier times, it was associated with progressive, severe deformities such as the so-called shepherd's crook deformity of the proximal femur or the so-called parrot's beak deformity of the femoral neck.

Like the radiographic variations at presentation, the histologic findings of fibrous dysplasia also may demonstrate an array of features. Some lesions may contain large amounts of benign cartilage, while others may have large cystic components. Histologically, the typical features include dysplastic truncated trabecular bone producing short, irregular bone segments (so-called Chinese alphabet soup). Dysplastic trabeculae typically are present within the fibrous stroma that replaces the normal bone, and the cellular activity of these lesions is moderate.

Treatment Treatment of fibrous dysplasia remains a challenging task in both children and adults. The typical patient who has a large, painful monostotic or polyostotic lesion usually benefits from intramedullary fixation of that osseous segment. A biopsy is required in most situations. Curettage is associated with a very high rate of local recurrence and therefore, in our experience, usually is not recommended. Thus, the lesions of fibrous dysplasia typically are best treated with biopsy followed by some type of cortical grafting or implant fixation to stabilize the long-bone segment. In the femur, this is

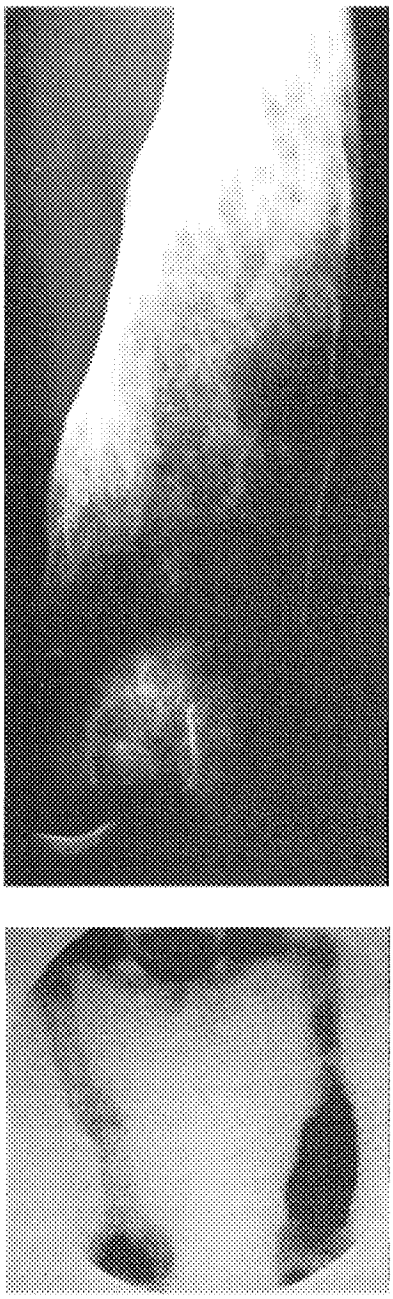

Fig. 16 Top, Radiograph of a skeletally immature individual with a long-standing history of a lesion in the distal left femur. Note the ground-glass appearance and the irregular, thinned cortex. **Bottom,** Bone scan showing dramatically increased uptake in the area of the lesion. This is a typical finding in active fibrous dysplasia, regardless of the age of the patient.

achieved with either a cortical fibular graft or a locking femoral nail, depending on the age of the patient. The child with a large lesion will have progressive, ongoing painful deformity, and multiple interventions may be required. This is especially true for patients with severe disease.

The overall prognosis for a patient who has fibrous dysplasia depends on the severity of involvement, which is related both to the involvement of individual bones and to the number of lesion sites.[50,51] The prognosis is good for a typical child who has a monostotic lesion, as the pain usually can be mitigated with prophylactic internal fixation. While the development of secondary osteosarcoma in a fibrous dysplastic segment of bone has been reported in the literature, it is an unusual problem.[4,51] The development of osteosarcoma from fibrous dysplasia is demonstrated radiographically by progressive erosion of bone or clinically by a soft-tissue mass, or both, and by increasing bone pain. The method of treatment should be selected carefully according to the severity of involvement and the radiographic and clinical findings for each patient.

Aneurysmal Bone Cyst

The typical aneurysmal bone cyst is an expansile, erosive cyst of bone that has a fairly high rate of local recurrence.[52] It occurs throughout childhood, with a slightly higher prevalence in adolescents than in pre-adolescents. It is unusual in patients in the third decade of life or older. The common anatomic sites of involvement include the proximal humerus, the femur, the tibia, and the ilium. The radiographic hallmark of aneurysmal bone cyst is a rapidly expansile cyst of bone that produces impressive thinning of cortices (Fig. 17). The lesion may well encroach on the growth plate. It may produce impressive inflammation in the surrounding bone, and in some instances it may be difficult to differentiate from osteosarcoma. When a patient presents with a pathologic fracture, the tumor may be very difficult to eradicate.

Diagnosis The histologic features of aneurysmal bone cyst include a hemorrhagic tissue with cavernous spaces. Surrounding those spaces may be fibroblastic tissue, multinucleated giant cells, and small fragments of bone. The cavernous spaces themselves have a lining of compressed mesenchymal cells. The differential diagnosis includes simple bone cyst and low-grade sarcoma.[53,54] Histologically, the differential diagnosis can be very difficult. Careful evaluation of the stromal cells will help to distinguish these entities.

Treatment Treatment of aneurysmal bone cyst includes curettage and bone grafting.[54] Local adjuvants, such as freezing with liquid nitrogen and application of phenol, have been promoted at some centers. The rate of local recurrence is high (44% [28 of 64] in one series),[52] and multiple recurrences may threaten an adjacent growth plate or joint surface and eventually necessitate more extensive resection. A preoperative bone scan may be a useful tool in assessing the degree of activity within a lesion that appears invasive radiographically. Alternative forms of treatment include injection of steroids or resection (for multiple recurrences).[3,4]

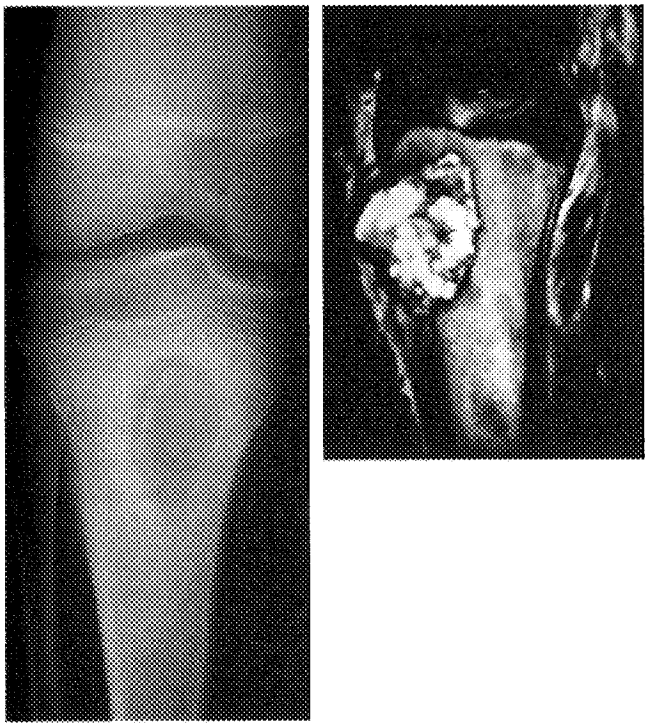

Fig. 17 Left, Plain radiograph showing erosive lucency in the proximal tibia of a 14-year-old child with a rapidly expansile aneurysmal bone cyst. **Right,** Sagittal MRI of the proximal tibia, showing inflammation and a large soft-tissue mass consistent with aneurysmal bone cyst.

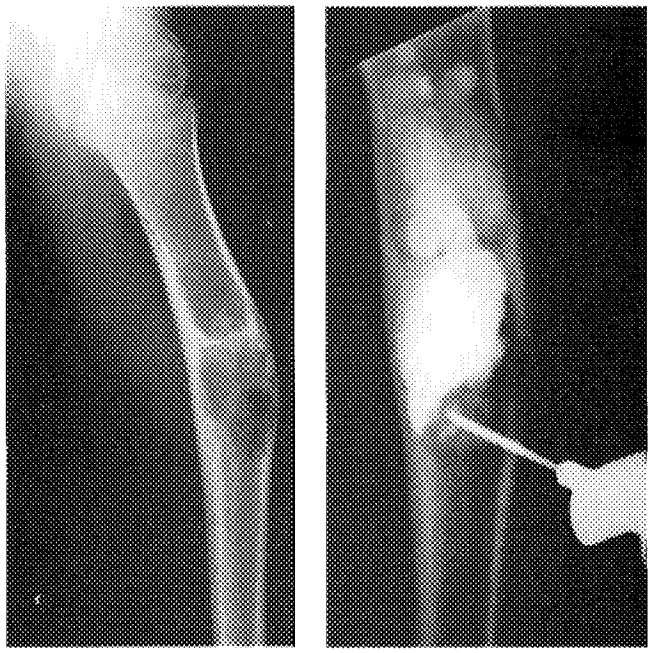

Fig. 18 Left, Lateral radiograph of the proximal humerus, showing a cystic lesion consistent with a unicameral bone cyst. **Right,** The injection of contrast medium into the lesion, showing containment in a bicameral lesion.

Unicameral Bone Cyst

Diagnosis Unicameral (simple) bone cyst has substantial overlap—histologically and radiographically—with aneurysmal bone cyst. The lesion may occur in either decade of childhood, and it occasionally occurs in adulthood. The male-to-female ratio is approximately 1:1. The most common site of involvement is the humerus, with almost 67% (277 of 416) of the lesions in one study occurring there.[53] Patients typically present with pain, with or without fracture.[53,55] Radiographically, unicameral bone cyst is a large cystic lesion with or without cortical thinning (Fig. 18). The lesion typically has been associated with a so-called fallen fragment sign (in which a piece of cortical bone has fallen to the bottom of the cyst), and it has a high risk of concomitant fracture. The histologic features of unicameral bone cyst typically include a space with a membrane composed of a thin rim of fibrous connective tissue. There may be a few multinucleated giant cells associated with this membrane, but the typical cyst has a very thin membrane, without the thickened trabecular spaces and abundant tissue that are present in an aneurysmal bone cyst.

Treatment Treatment alternatives for unicameral bone cyst primarily include the injection of steroids or curettage and bone grafting. Despite the good early results that have been achieved with the injection of steroids, that technique has been associated with local recurrence and persistent radiolucencies.[53,55] Curettage and bone grafting is an alternative form of treatment, as is curettage combined with freezing with liquid nitrogen or application of phenol. In one study, the rate of local recurrence following surgical curettage was 14% (25 of 178), while the rate of failure after the injection of steroids was 24% (34 of 141).[53] Some unicameral bone cysts have a risk of multiple recurrence and present a challenge for local control. A patient who has a pathologic fracture probably has a more aggressive lesion and a poorer prognosis.

Eosinophilic Granuloma and Histiocytosis X

Histiocytosis X, or Langerhans-cell granulomatosis, describes a spectrum of disease involvement that has been characterized by three syndromes: Letterer-Siwe syndrome, Hand-Schüller-Christian syndrome, and eosinophilic granuloma.[56] All of these syndromes involve proliferation of histiocytes. Symptoms associated with these syndromes vary according to the various systems involved. Letterer-Siwe disease is more severe and is seen in younger patients who present with hepatosplenomegaly, pulmonary disease, and anemia, with or without dermatitis. These children usually are severely ill and may be immune deficient. The

disease progresses along a malignant course that may necessitate chemotherapy. The long-term prognosis is poor.

Hand-Schüller-Christian disease generally involves children between the ages of 3 and 12 years.[3] These children may present with hepatosplenomegaly, exophthalmus, diabetes insipidus, mastoiditis, and dermatitis. A small percentage of these children also may have a malignant course over a longer period of time. More commonly, they have a less aggressive form of the disease. One of the bigger problems is diabetes insipidus, which is extremely difficult (if not impossible) to reverse and is best detected early.

Eosinophilic granuloma is a more limited disease process. Usually, it is characterized by a solitary bone lesion. Children who have eosinophilic granuloma do, however, require careful staging to rule out other sites of involvement. Appropriate staging includes a total-body technetium bone scan or skeletal series, blood work (complete and differential blood-cell counts and tests for liver enzymes), and radiographs of the skull and pelvis. Children who have multisystem disease require more aggressive management, including steroid-based chemotherapy.

By far the most common anatomic site of involvement of histiocytosis is the skull (Fig. 19, *left*). Other osseous sites include the pelvis, the proximal femur, the spinal column, the ribs, the hands, and the feet. Classic involvement of the spine with vertebral collapse is referred to as vertebra plana and has a coin-on-edge appearance (Fig. 19, *right*).

Diagnosis The radiographic picture of eosinophilic granuloma is primarily a cystic lesion of bone, but with a very inflamed, aggressive-appearing reaction to the lesion.[57] It truly is the great imitator among pediatric bone tumors and, like osteomyelitis, should be included in all differential diagnoses in children, especially those who are less than 12 years old. The dominant radiographic hallmark is a permeative lesion associated with a lucency. In the skull, it may present a picture of punched-out lesions, which may be quite large. In the long bones, the osseous reaction is impressive; however, typically there is not a large soft-tissue mass associated with this lesion, and this helps to distinguish it from malignant tumors of bone such as Ewing's sarcoma.

Histologically, there is dominance of proliferative histiocytes. Eosinophils also are quite prominent. This round-cell histologic appearance can be quite difficult to distinguish from that of other inflammatory conditions such as osteomyelitis, from round-cell tumors such as Ewing's sarcoma, and from primitive neuroectodermal tumor, especially on frozen section. Immunohistochemical techniques may be helpful in excluding other diagnoses.[3]

Treatment The treatment of eosinophilic granuloma includes biopsy and curettage, with or without grafting.[3,6] If a complete curettage is carried out, the lesion rarely recurs. Injection of cortisone also has been attempted.[58] An alternative form of treatment for a patient who has an eosinophilic granuloma in an inaccessible site is the use of low doses (400 to 1000 rads [centigrays]) of radiation. Radiation therapy usually is reserved for lesions in the spine, the skull, and, occasionally, the sacrum and the pelvis.[58] Overall, the reported rate of survival for Letterer-Siwe syndrome is

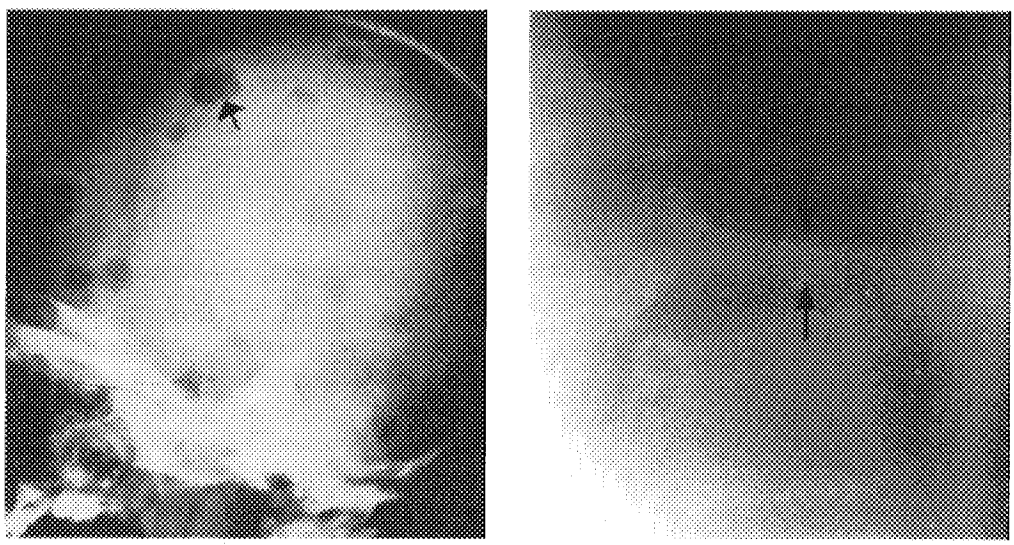

Fig. 19 **Left,** Lateral view of the skull of a 2-year-old child showing multiple defects (arrow) in the lateral aspect of the skull consistent with polyostotic eosinophilic granuloma. **Right,** Vertebra plana (arrows) of the first lumbar vertebra. This is a typical pattern of histiocytosis involving a vertebral body.

low and that for Hand-Schüller-Christian syndrome is 90%.[6] In our experience, the rate of survival for eosinophilic granuloma is virtually 100%. The prognosis for any particular child is related to the extent of involvement rather than the age at presentation.

Conclusion

The treatment of benign bone tumors needs to be individualized on the basis of the known natural history of the lesion and its biologic behavior. There are several potential pitfalls, but they can be minimized with a careful systematic approach to these tumors. When in doubt, however, consultation with an experienced orthopaedic oncologist will help to determine the best approach for the patient. Doing so will minimize the risk of undertreatment or overtreatment.

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